

LEADLESS PACEMAKER IMPLANTATION VIA THE INTERNAL JUGULAR VEIN IN A 10-YEAR-OLD CHILD WITH KEARNS-SAYRE SYNDROME

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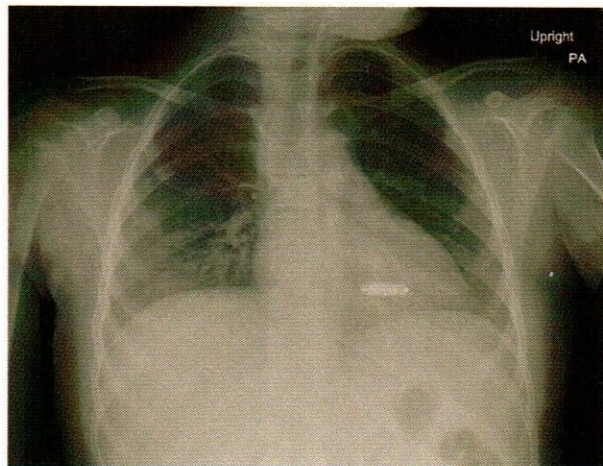
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Introduction:

Kearns-Sayre syndrome (KSS) is a rare mitochondrial disorder; frequently with cardiac manifestations. The cardiac system is of particular significance as patients with KSS may progress to complete heart block. Prophylactic pacemaker implantation can therefore lengthen life expectancy in KSS. Leadless pacemaker implantation offers a minimally invasive alternative to traditional pacemakers; particularly in those with challenging anatomy. In this presentation, we report a pediatric patient with leadless pacemaker implantation for KSS.

Case report:

A female with KSS presented at age 7 for evaluation of cardiac involvement related to her mitochondrial disease. Her past medical history included pigmentary retinopathy, sensorineural hearing loss, failure to thrive, renal tubular disorder, carnitine deficiency, hypomagnesemia, and global developmental delay. Initially, her cardiac evaluation was normal. At age 9, her EKG showed right axis deviation, non-specific intraventricular conduction delay and borderline long QTc. A few months later during an elective surgical procedure she developed wide-complex tachycardia, from which she self-converted to normal sinus rhythm. At that time Holter monitoring showed sinus rhythm with intermittent right bundle branch block, without pauses. At age 10, she was diagnosed with diabetes mellitus type 1. Her echocardiogram continued to show normal chamber sizes and heart function. Her EKG demonstrated sinus rhythm with 1st degree AV block, right bundle branch block, and a QTc of 483. Holter monitoring showed she had progressed to a high grade (2:1) AV block with a 1.4 second pause. The decision was then made to place a prophylactic pacemaker. However, she was notably a poor candidate for traditional transvenous pacemaker placement due to her small, stenotic subclavian veins bilaterally. Therefore, a



leadless pacemaker was placed through the internal jugular vein. She did well and was discharged the day after placement. Her cardiac status remains stable one year later with ventricular pacing at 0.5%.

Discussion:

Treatment for KSS is primarily supportive. Some patients with KSS die suddenly, likely from progression to complete heart block. Therefore, pacemaker placement at the first sign of conduction changes on EKG ought to be discussed due to the unpredictable natural progression to complete AV block. A leadless pacemaker should be considered for patients requiring pacemaker placement as an alternative to traditional pacemakers; particularly in the challenging anatomical patient.